Extracranial temporal cavernous hemangioma: Differential diagnosis, and a review of literature

Sir,
A small swelling in the temporal region is often benign and managed conservatively unless it causes local pain, shows an increase in size or becomes cosmetically disfiguring. The differential diagnosis of pathology in this region is quite varied and includes soft tissue tumors such as lipoma, schwannoma, dermoid, mesenchymal angiolipoma, and spindle cell hemangioendothelioma; vascular malformations such as cavernous hemangiomas and arterio-venous malformations (AVMs); and malignant lesions such as liposarcomas and angiosarcomas. Occasionally, extracranial extensions of intracranial meningiomas/hemangiopericytomas or osteoma of the calvarium can present in a similar fashion. Enlarged lymph nodes, myositis ossificans, and temporal arteritis complete the list.

We discuss the case of a 61-year-old female patient with an intramuscular cavernous hemangioma of the temporalis muscle and review varying presentations and the differential
diagnosis. Due their rarity and vague presentations, this entity presents significant diagnostic challenges.[1]

This 61-year-old female patient presented with a small swelling in the left temporal region of her head 4 years previously. She was initially asymptomatic and her family physician, who suspected that this was a lipoma, recommended a conservative approach. More recently, she noticed a significant growth in the size of the lesion over 1 year, which prompted a referral. Apart from the increasing size of the lesion, she was otherwise asymptomatic. Examination revealed a smooth, nonpulsatile, nontender, immobile globular swelling in the left temporal region. It was firm and noncompressible. There was no audible bruit. On examination, she had no neurological deficit.

Magnetic resonance imaging (MRI) revealed a 3.5 cm × 2.5 cm × 4.5 cm lesion within the left temporalis muscle. There was no involvement of the underlying bone or scalp. The lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images and showed heterogeneous enhancement with administration of gadolinium [Figure 1]. There was neither restriction of diffusion on the diffusion-weighted imaging nor was there evidence of hemorrhage on susceptibility-weighted imaging [Figure 2]. An angiogram ruled out an AVM, but showed evidence of minimal neovascularity of the mass. There were two small areas of contrast blush in the superior and inferior aspect of the lesion that appeared in the arterial phase and persisted, slightly enlarging into the venous phase. However, the bulk of the lesion did not demonstrate a vascular blush [Figure 3]. The preoperative diagnosis was that of a cavernous hemangioma. Surgery was recommended both for the confirmation of diagnosis as well as cosmesis. A frontal-temporal skin flap was turned forward to the orbital rim. The tumor was then seen and felt within the temporalis muscle, from which it was dissected en bloc, excising it radically. There was no infratemporal or orbital extension.

Postoperatively, the patient developed a left frontalis paresis that had significantly improved at 4 weeks. The postoperative computed tomography (CT) scan done showed no residual lesion [Figure 4].

Histopathology showed a proliferation of large, thick-walled vascular channels with foci of lymphoid inflammation and pieces of striated muscle. This was consistent with a cavernous hemangioma [Figure 5].

Less than 1% of hemangiomas are intramuscular and only 14% occur in the head and neck.[1-4] The masseter and trapezius muscles are the most frequently affected sites.[2,4] Involvement of the orbicularis oris, mentalis, digastric and mylohyoid muscles has also been reported.[5] Intramuscular hemangiomas within the temporalis muscle are extremely rare. Histologically they are classified as capillary (small vessel), cavernous (large vessel), and mixed, depending on the predominant vascular channels that they are composed of.[6] Minor trauma, hormonal status, or excessive muscle contraction is believed to play a role in their growth by inducing changes in the blood flow. They have been reported to grow to a giant size, cross tissue planes, and can rarely have an intracranial extension, producing a mass effect on the brain.[7] Table 1 discusses the clinical, radiological, and pathological differential diagnosis of these lesions within the temporalis muscle.

While in most patients they present as a painless, progressive swelling within the temporalis muscle causing facial asymmetry, there are reports of cases presenting with significant pain and temporo-mandibular joint dysfunction.[8] They can change in size with a Valsalva maneuver due to large venous channels within. They are also known to occasionally develop several years after blunt trauma to the head. The diagnosis is more straightforward when there is an associated cutaneous change, but this is uncommon.
While classically described as soft and compressible, cavernous hemangiomas can occasionally be firm and mistaken for soft tissue tumors. Rarely, they can be pulsatile and may have a bruit mimicking an AVM. Itosaka et al. have described a case where repeated hemorrhages within the tumor resulted in spontaneous regression of the hemangioma. While a long-standing history of a slowly growing painless mass suggests a benign pathology, a short history of a rapidly growing lesion could signal something more sinister and should not be managed expectantly.
MRI is the investigation of choice in defining the nature of the tumor and in differentiating its vascular nature from soft tissue elements. These lesions are most often well-circumscribed within the muscle, isointense on T1-weighted images, and hyperintense on T2-weighted images. Occasionally, a hypointense rim similar to what is seen in cavernous malformations of the brain is also visible on the T2-weighted images. Some authors suggest that a serpiginous pattern on T2-weighted images is characteristic. The heterogeneity on enhancement is due to thrombosis, hemosiderin deposition, and calcification. Phleboliths can sometimes be seen inside the lesion as low-intensity areas. Lipomas, angiolipomas, and dermoids are usually hyperintense on T1W images, whereas schwannomas and meningiomas have a more uniform enhancement. A CT scan may show remodeling of the underlying skull in long-standing cases.

Angiography is helpful to differentiate cavernous hemangiomas from an AVM. AVMs will have either a compact or a diffuse intramuscular nidus and the presence of early draining veins. The main arterial feeders are branches of the superficial temporal and internal maxillary artery, and the draining veins are commonly the facial and external jugular. Cavernous malformations may be angiographically occult (involuting type) or they may show uniform hypervascularity (proliferating type). There may be separate visible areas of blush due to pooling of stagnant blood, as seen in our case. Angiolipomas, on the other hand, show coarse neovascularity and mottled staining during the capillary phase. These findings should guide in the diagnosis, avoiding the need for an open biopsy, which could lead to unwarranted hemorrhage.

Hemangiomas are benign and not known to metastasize, but can have local recurrence due to infiltration of muscle and soft tissue. Capillary hemangiomas are more aggressive than the cavernous and mixed types. The objective of surgery should be total removal, preferably in an en bloc fashion. Care should be taken to prevent injury to the temporal and auricular branches of the facial nerve because if permanent, this complication could be more disfiguring than the swelling itself. Ohyama et al. used an electric nerve stimulator where the facial nerve ran over the skin as well as subfacially to avoid this complication. Steroids, sclerosing agents, and radiation are other proposed treatment modalities but are not consistently effective.

Due to the wide spectrum of diagnostic possibilities, a soft tissue swelling in the temporal region requires careful evaluation with an MRI as well as angiography, as the diagnosis has a significant impact on the management of these lesions. The definitive diagnosis is usually made by histopathological evaluation. Careful surgical planning and total removal usually result in a good result.

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References

Pediatric intraparenchymal meningioma: A review of literature

Sir,

Meningiomas are rare tumors in the pediatric population accounting for approximately 1 to 2% of all intracranial neoplasms in this age group. [1] An intraparenchymal meningioma, which consists of a meningioma without a dural tail and is surrounded by the normal brain parenchyma is very rare, with very few case reports available in the literature. [2]

We report one such case and present a pertinent literature review. A 16-year-old male child presented with a history of multiple seizures. The last seizure occurred 4 months back, and the patient was seizure free after starting oral phenytoin tablets. Patient had no neurological deficits on examination. Magnetic resonance imaging (MRI) revealed a tumor, which was hypointense on T1-weighted image (T1WI), iso to hypointense on T2WI, present in the right cingulate and superior frontal gyrus. There were areas of blooming on susceptibility weighted imaging (SWI), which corresponded to the areas of calcification on noncontrast computed tomography (CT) scan of the head. A heterogenous enhancement was seen on post-contrast scan [Figure 1]. Considering the radiology, a preoperative diagnosis of oligodendroglioma was made.

The patient underwent a right frontal craniotomy, an interhemispheric approach, and excision of the tumor. The tumor was present in the right medial frontal lobe surrounded by normal brain parenchyma. Resection of the overlying cortex was done to approach the tumor. The superficial part of the tumor was firm and fibrous, and calcification was present in the deeper portion of the tumor. A branch of the anterior cerebral artery (ACA), which was adjacent to the tumor, was injured during the surgical dissection, as there was a very poor plane of cleavage between the tumor and the vessel; the injured vessel was secured with ligacips. Circumferential dissection around rest of the tumor did not reveal any attachment to the dura. The part of the tumor that was densely adherent to the tumor was left behind to preserve the vessel.

[Figure 1: (a) Non-contrast computed tomography (NCCT) of the head showing the tumor in the right medial frontal lobe with calcification. (b) T1-weighted image (T1WI) axial section showing the hypointense tumor within the cortex. (c) T2WI axial section showing the hypointense tumor in close proximity to the anterior cerebral artery without any perilesional edema. (d and e) Contrast enhanced images, axial and coronal section, showing the heterogeneous enhancement in the tumor. (f) Susceptibility weighted imaging showing blooming in areas corresponding to the calcified areas on plain CT scan of the head]

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